Radiology Corner

Sinus Venosus Atrial Septal Defect

Guarantor: MAJ Robert A. Jesinger, USAF, MC[*]
Contributors: MAJ Robert A. Jesinger, USAF, MC[*]; Diane C. Strollo, MD, FCCR[†]

Note: This is the full text version of the radiology corner question published in the March 2010 issue, with the abbreviated answer in the April 2010 issue.

We present a case of sinus venosus atrial septal defect in a patient who sustained trauma. A 58-year-old man presented to the emergency department (ED) with chest pain and an ankle fracture after being hit by a car while riding a horse. Chest imaging noted enlarged central pulmonary vascularity. Chest CT revealed partial anomalous right pulmonary venous return to the superior vena cava and right atrium and sinus venosus atrial septal defect. Transesophageal echocardiogram confirmed these findings as well as identifying Eisenmenger's physiology. In this full-text version, we present a more detailed discussion of sinus-venosus atrial septal defect associated with partial anomalous pulmonary venous return.

Summary of Imaging Findings

A 58-year-old male was riding a horse when they were both struck by a motor vehicle. The man was thrown approximately 5 to 8 feet and suffered open comminuted fracture of the left ankle. On evaluation in the emergency department, the patient complained of chest and leg pain. His respirations were 24 per minute and his oxygen saturation on room air was 98%. His vital signs were otherwise unremarkable. A portable chest radiograph was obtained (figure 1). The patient was admitted and underwent orthopedic repair of his ankle fracture. Three days later, he developed dyspnea and non-specific chest pain. He reported symptoms of progressive shortness of breath over several years. A contrast-enhanced chest CT was performed to evaluate for pulmonary embolism (figure 2).

The chest radiograph (figure 1) demonstrates increased central pulmonary vascularity ("pulmonary arterial hypertension" pattern) without evidence of pulmonary edema; the cardiac silhouette was questioned to be mildly enlarged, primarily from right heart enlargement.

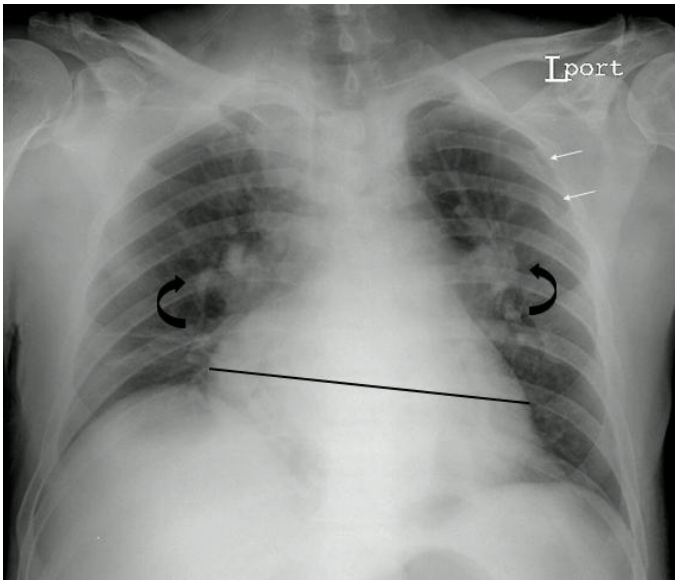

Fig. 1 AP Chest radiograph (CXR) demonstrating increased central pulmonary vascularity (curved black arrows) with mild cardiomegaly (black line). Old left 3rd and 4th rib fractures are noted (straight white arrows).

Contrast-enhanced chest CT (figure 2) demonstrated two anomalous pulmonary veins: one draining into the superior vena cava and the other into the right atrium. A communication between the superior aspect of the right and left atria was identified.

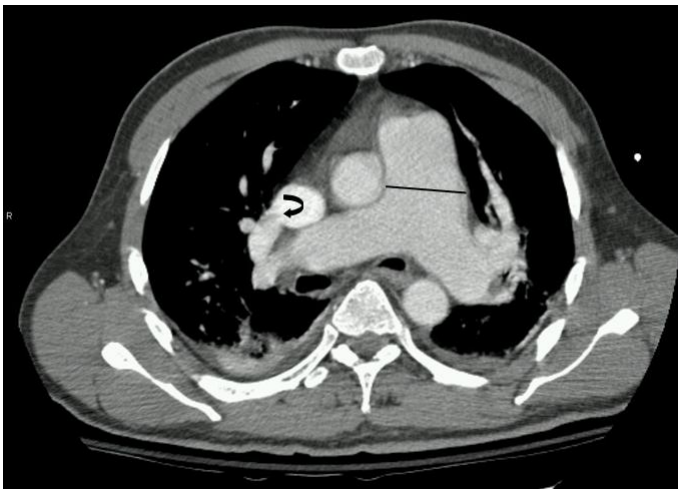

Fig. 2A Contrast-enhanced chest CT demonstrating an anomalous pulmonary vein draining into the superior vena cava (black curved arrow). Marked main pulmonary artery enlargement is also noted (black line) as the diameter of the main pulmonary is 50% greater than the ascending aorta (normally similar in size).

[*] Department of Radiology (60 MDOS); David Grant USAF Medical Center, Travis AFB, CA 94535.
[†] Department of Radiology; University of Pittsburgh Medical Center, Pittsburgh, PA 13215.
[‡] Department of Radiology and Radiological Sciences; Uniformed Services University of the Health Sciences, Bethesda, Maryland 20814-4799

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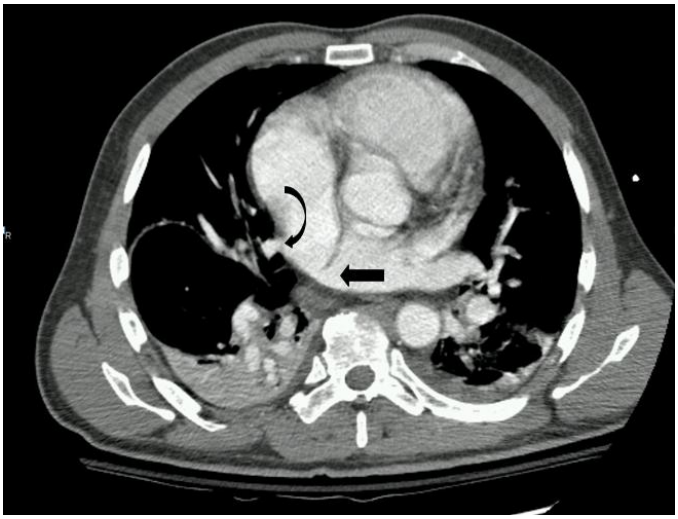

Fig. 2B Contrast-enhanced chest CT demonstrating a communication between the superior right and left atria (straight black arrow). A second anomalous pulmonary vein drains into the right atrium (curved black arrow).

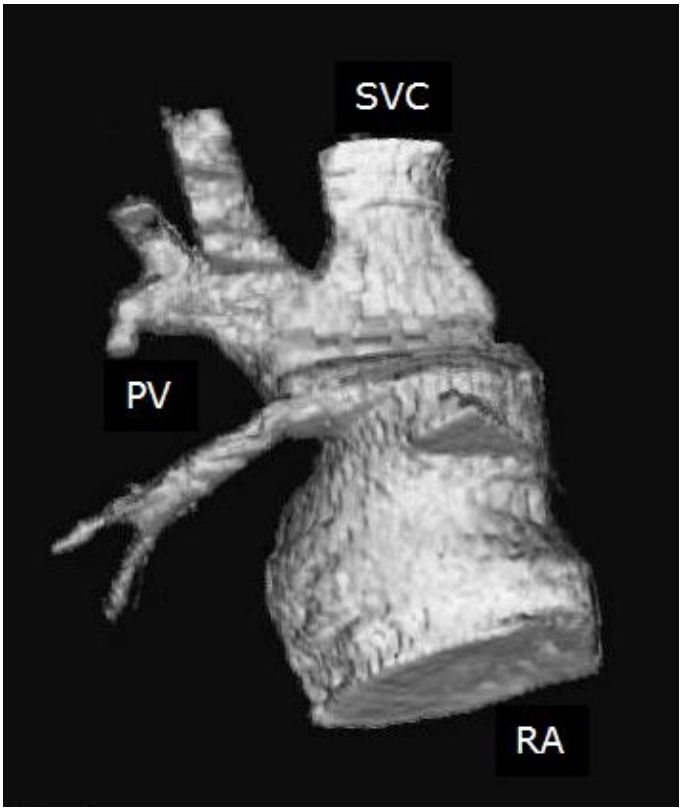

Fig. 2C Chest CT reconstruction demonstrating the anomalous pulmonary veins draining into the superior vena cava (SVC) and right atrium (RA).

Transesophageal echocardiogram (figure 3) demonstrated right heart enlargement with a right-to-left shunt through the communication between the superior left and right atria. Color Doppler images demonstrate a right-to-left shunt.

Fig. 3A Transesophageal echocardiogram (TEE) demonstrating right heart enlargement.

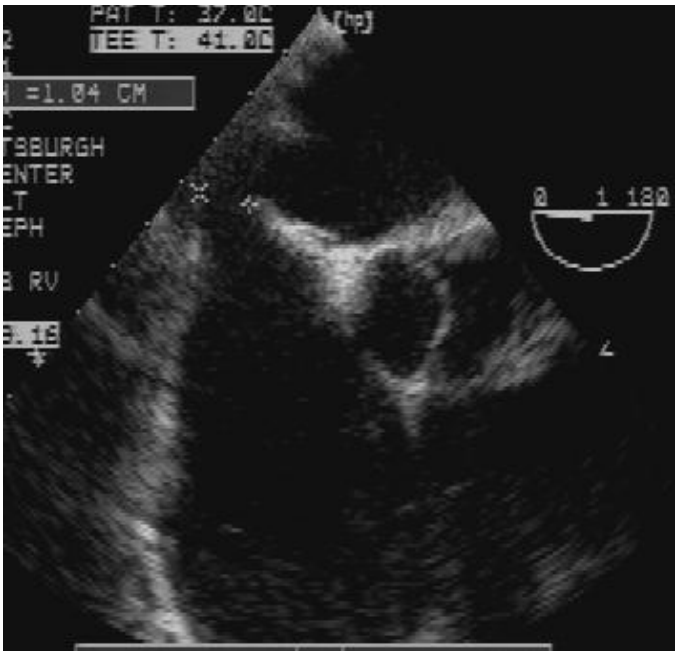

Fig. 3B Transesophageal echocardiogram (TEE) demonstrating a sinus venous defect measuring 1cm in size.

Diagnosis:

Sinus Venosus Atrial Septal Defect

The patient's TEE and cardiac catheterization confirmed sinus venous atrial septal defect with bidirectional flow (predominantly right-to-left) suggesting Eisenmenger's physiology. The right heart was moderately enlarged with

right ventricular hypertrophy and pulmonary arterial hypertension (pulmonary arterial pressures were 60/25 mmHg). The patient underwent bovine pericardial baffle patch repair of the sinus venosus defect with re-implantation of the anomalous right pulmonary veins into the left atrium. At surgery, the interatrial defect measured almost 3cm. The right superior pulmonary vein drained anomalously into the posterior aspect of the superior cavo-atrial junction, and the right middle lobe segmental veins drained separately into the right atrium (as demonstrated in figure 2C). The right inferior pulmonary vein and left pulmonary veins drained normally into the left atrium. The patient tolerated his surgery well and recovered uneventfully.

Fig. 3C Transesophageal echocardiogram (TEE) color Doppler demonstrating a right-to-left shunt between the right and left atria.

Discussion

This case demonstrates the classic findings of sinus venosus atrial septal defect (ASD), a rare congenital deficiency of the superior inter-atrial septum typically associated with partially anomalous right pulmonary venous return (PAPVR) to the superior vena cava and right atrium. The sinus venosus defect typically results in a left-to-right shunt at the level of the atria. The presence of PAPVR compounds the problem with additional left-to-right shunt pathways.

While the exact embryology is controversial, the sinus venousus defect may actually represent an "unroofing" defect of the right pulmonary veins at the superior aspect of the interatrial septum rather than a true deficiency of the atrial septum.[1] The sinus venosus defect is typically classified with the atrial septal defects (ASD), as both may result in a left-to-right shunt. The two main types of ASD are ostium primum and ostium secundum defects (table 1).[2]

Defect	Incidence	Characteristic Defect Location
Ostium primum	15 %	Inferior
Ostium secundum	75 %	Mid-septal
Sinus venosus	10 %	Type I: junction of SVC and RA Type II: junction of IVC and RA

Table. 1 Classification of atrial septal and sinus venosus defects.

Sinus venosus defects are thought to occur during the development of the pulmonary veins during the fourth to seventh weeks of embryogenesis (figure 4).[3]

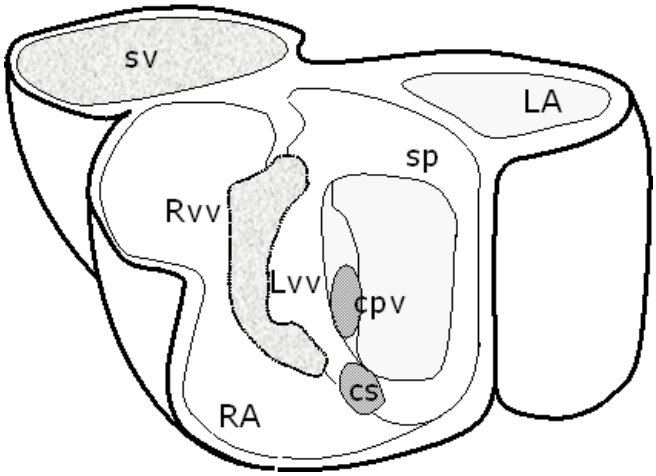

Fig. 4 Pulmonary venous development at 5 weeks gestation. [cpv=common pulmonary vein, cs=coronary sinus, LA=left atrium, Lvv=left venous valve, RA=right atrium, Rvv=right venous valve (future crista terminalis), sv=sinus venosus, sp=septum primum]. (drawing by Maj Robert Jesinger)

The sinus venosus is the most common venous receptacle that drains into the primitive right atrium of the embryonic heart. During development, the common pulmonary vein (CPV), which originates within the sinus venosus, is repositioned into the left atrium as the sinus venosus normally blends into the posterior walls of the atria. The common wall of the CPV and proximal right superior pulmonary vein contributes to the superior portion of the atrial septum (figure 5).[4]

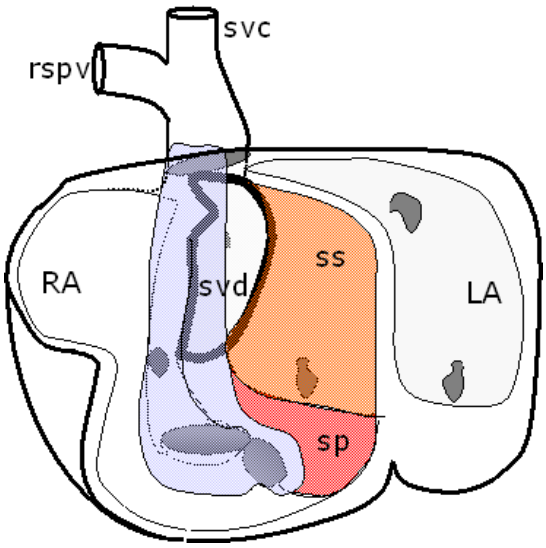

Fig. 5 Sinus venosus defect at birth. The shaded area in purple represents the sinus venosum. The anomalous right pulmonary venous anatomy shown was seen in our case. [LA=left atrium, RA=right atrium, rspv=right superior pulmonary vein, sp=septum primum, ss=septum secundum, svc=superior vena cava, svd=sinus venosus defect]. (drawing by Maj Robert Jesinger)

A deficiency of this common wall can result in a superior interatrial communication (Type I) as well as anomalous pulmonary venous drainage to the right atrium or SVC due to "unroofing" defects.[1,5] Rarely, the sinus venosus defect may arise inferiorly (Type II), at the orifice of the right inferior pulmonary vein, IVC, and right atrium. In either case, these defects do not typically interfere with the concurrent development of the atrial muscular septum primum or septum secundum. Occasionally, partially anomalous left pulmonary venous return (PAPVR) to the superior vena cava may be seen (figure 6).

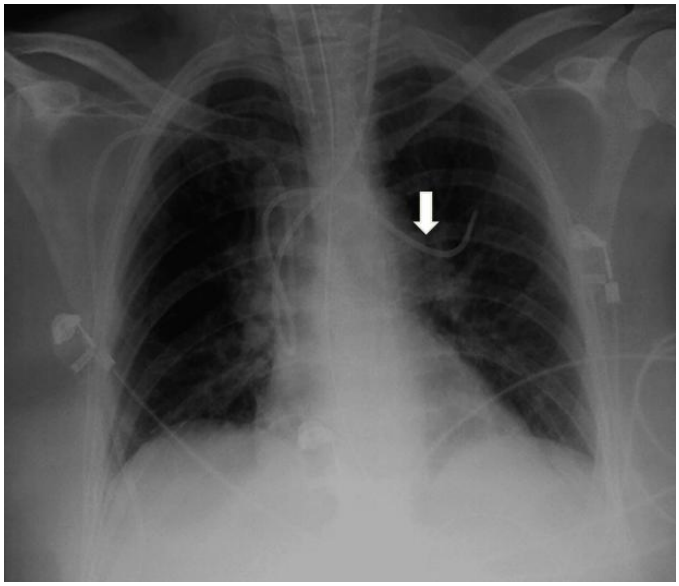

Fig. 6 AP Chest radiograph (CXR) demonstrating a Swan catheter (placed via the left internal jugular vein) looped in the SVC and coursing into an anomalously draining left upper lobe pulmonary vein (white arrow).

ASD comprises approximately 10% of congenital heart disease at birth and 25% of adult congenital heart disease. Next to bicuspid aortic valve, ASD is the second most common cause of adult congenital heart disease.[2]

Most adult cases of ASD, including sinus venosus defects, remain unrecognized until the fourth to fifth decades of life when pulmonary arterial hypertension and right-to-left shunting develops.[6] Consequently, patients with ASD are at risk for developing atrial dysrhythmias with an inherent risk of thromboembolic complications. Sinus venosus defect may carry a worse prognosis than other forms of ASD and may need to be treated at a younger age, via medical therapy and/or surgical closure. Surgical repair of ASD, including sinus venosus defect, in patients over 40 years of age, increases long-term survival and decreases the incidence of heart failure.[7,8]

Summary

Atrial septal defects can present with pulmonary arterial hypertension in adults. With the increased use of CT, careful assessment for these defects, and their associated pulmonary venous anomalies in patients with enlarged central pulmonary vessels, may be beneficial as surgical repair of these defects can improve outcomes.

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References

1. Van Praagh S et al. Sinus venosus defects: unroofing of the right pulmonary veins – anatomic and echocardiographic findings and surgical treatment. Am Heart Journal 1994; 128: 365-79.
2. Miller SW. Cardiac Imaging: the Requisites, 2nd ed. Elsevier Mosby 2005: 36-38; 324-325.
3. Blom NA et. al. Normal development of the pulmonary veins in human embryos and formulation of a morphogenetic concept for sinus venosus defects. Am J Cardiology 2001; 87:305-09.
4. Sadley TW. Langman's Medical Embryology, 7th ed. Williams and Wilkins 1995: 191-201.
5. Al Zaghal AM et. al. Anatomic criteria for the diagnosis of sinus venosus defects. Heart 1997; 78: 298-304.
6. Vogel M, Berger F, Kramer A, et. al. Incidence of secondary pulmonary hypertension in adults with atrial septal or sinus venosus defects. Heart 1999; 82: 30-33.
7. Agrawal SK, Khanna SK, Tampe D. Sinus venosis atrial septal defects: Surgical follow-up. European J Cardiothoracic Surg 1997; 11(3): 455-7.
8. Konstantinides S et. al. A comparison of surgical and medical therapy for atrial septal defects in adults. NEJM 1995; 333(8): 469-73.